904-47 Permanent Overdrive Atrial Pacing in Chronic Management of Atrial Reentrant Tachycardias in Postoperative Patients with Congenital Heart Disease

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Sustained atrial tachycardias occurring after surgery for congenital heart disease, are often difficult to control by antiarrhythmic drugs and by antitachycardia pacing. We implanted a permanent pacing system in 11 pts with drug resistant recurrent atrial reentrant tachycardias. 7 pts were male and 4 female; mean age was 7.4 years (range 1 to 28 yrs); mean time from surgery to the first documented atrial tachycardia was 2.2 years (range 1 month to 9 yrs). All pts had been unsuccessfully treated with two to five antiarrhythmic drugs before pacing. At implantation the atrial pacing rate was programmed to be 20% faster than the mean diurnal spontaneous rate. Individualized programs of pacemaker rate settings were then selected following serial 24 hours Holter monitoring, to obtain a prevailing paced atrial rhythm. Mean follow-up was 3.4 years (range 6 mos to 5 yrs). 3 pts had recurrences of atrial tachycardia during the first six months after the implantation, 8 pts remained arrhythmia free. No patient had late recurrences and only two are still on antiarrhythmic drug therapy. *In conclusion:* permanent overdrive atrial pacing is a very useful tool in chronic management of postoperative atrial reentrant tachycardias.

904-48 Risk Factors for Venous Obstruction in Children with Transvenous Pacing Leads

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To determine the incidence and risk factors for venous obstruction (OBST), we prospectively evaluated with echocardiography 63 of 70 eligible children who had transvenous pacing leads placed between 1985 and 1993. The median (range) age at initial implantation was 7.6 yrs (0.7, 16), and 8 patients had subsequent additional implants. OBST was defined as a combination of Doppler flow abnormalities in the SVC or innominate (InnV) vein and a 2D echo appearance of vessel narrowing and/or the clinical appearance of dilated superficial veins. OBST was noted in 13/63 (21%) patients, with location of OBST at the distal subclavian vein in 5, SVC in 4, InnV-SVC junction in 2, and multiple sites in 2. Venography in 11 of these patients (2 refused) showed that the severity of OBST (as defined by % luminal narrowing) was complete (100%) in 3 patients, severe (>90%) in 4, and moderate (60–90%) in 4. Of the 8 patients who had additional implants, 3 (38%) had OBST. Risk factors for OBST in the remaining 55 single implant patients (10 with OBST; 18%) were explored. Patients with vs. without OBST did not differ regarding date or duration of implant, number of leads, lead material or the presence of associated heart defects or surgery. Patients with OBST were younger at implant (median 5.6 vs. 8.8 yrs; $p < 0.05$). Total cross-sectional area of lead(s) was related to body surface area at implant (RATIO). Patients with OBST had higher mean RATIO (7.6 ± 1.6 mm^2/m^2) than patients without OBST (4.9 ± 2.0 mm^2/m^2; $p < 0.0002$). After controlling for RATIO in multiple logistic regression, no other variable predicted OBST. Receiver-operator characteristic curves showed a RATIO of >6.6 mm^2/m^2 to best predict OBST, with a sensitivity of 90% and specificity of 84%.

Conclusion: Since pacing is lifelong, sizing of transvenous leads to the child is important to prevent OBST and preserve venous access.

904-49 Children Below 6 Years of Age with Accessory Pathway Related Tachyarrhythmias. Whom to Ablate and How

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By August 1994, a total of 103 children (≤15 yrs) underwent radio-frequency current (RFC) ablation for accessory pathway (AP) mediated supraventricular tachycardia (SVT). Of these, 23 were infants aged less than 6 years (11 f, 12 m; 3.9 ± 1.6 yrs). All infants had suffered from recurrent or almost incessant episodes of SVT, which were diagnosed at birth in 13 and prenatally in 4. SVT was refractory to an antiarrhythmic regimen with 3 to 6 different agents. Tachycardia-related symptoms included cardiac arrest in 2 infants with WPW, syncope in 3 pts, reduced physical stress capacity in 11 and SVT-related chest pain in 1. Thirteen pts had an overt AP, with an additional concealed pathway in 2; the remaining 10 pts had a concealed AP, in 6 of which it sustained the permanent form of junctional reciprocating tachycardia (PJRT). In the latter group 3 infants had reduced left ventricular function (Fraction-shortening 13 to 21). In 11 infants, a single 5- or 6 French steerable catheter was used for therapeutic intervention. In 9 infants a second catheter was required for diagnostic purposes. Within 25 sessions, 22 infants were cured by a median of 7 RFC applications. Procedure duration was 3.4 ± 1.9 hours, with a median radiation exposure time of 29.0 minutes. During a 19 month follow-up (median), 20/22 infants were asymptomatic and required no antiarrhythmic medication. Two pts had recurrences of SVT (1 PJRT), one of which underwent a successful repeat session. Tachycardia related reduced left ventricular function evolved to normal values in all 3 infants within the follow-up period. There were no serious complications.

Conclusions: 1) RFC-ablation of accessory pathway-mediated supraventricular tachycardia is feasible, safe and effective in children aged less than 6 years. 2) Reduced invasiveness due to simplified catheter techniques is achievable in this particular patient population. 3) In children with an age <6 years, indication for RFC-ablation is restrictive and should be provided in pts with severe symptoms due to drug-refractory SVT.

904-50 Pacemaker Cardioverter Defibrillators in Young Patients

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Although life-threatening ventricular dysrhythmias are uncommon in young patients, there are underlying heart diseases that may predispose these patients to ventricular tachycardia or ventricular fibrillation (VT\VF). To test the hypothesis that tiered therapy with a pacemaker-cardioverter defibrillator (PCD) is a therapeutic option for young patients at risk for sudden death, we reviewed our experience with these devices. Nine patients, aged 9–33 years (17.7 ± 7, mean ± SD) at the time of implantation, received PCD's between March, 1993, and the present. Post-operative diagnoses included complete repair of tetralogy of Fallot (2), Senning for D-transposition of the great arteries (1), double outlet right ventricle, status post ventricular septal defect closure baffling the left ventricle to the aorta (1), and Fontan for single ventricle with pulmonary stenosis (1). Other diagnoses included Wolff-Parkinson-White (WPW) (1), WPW and dilated cardiomyopathy (1), arrhythmogenic right ventricular dysplasia (1), and long QT syndrome (1). All patients had inducible or clinical unstable VT\VF before implantation except one patient with WPW who had a family history of sudden death. Seven of the patients had failed antiarrhythmics before implantation. Defibrillator electrode configurations consisted of epicardial patches (4), transvenous alone (3), and transvenous with subcutaneous patch(es) (2). In 10.4 ± 5 (range 2–17) months of follow-up, overdrive pacing was successful in one patient, and there have been appropriate defibrillator discharges in five patients (56%). Seven of the nine patients remain on antiarrhythmics. There was only one complication, a moderate pericardial effusion two months following transvenous lead implantation in a 24.9 kg nine-year old. No system has required revision to date.

Conclusion: In relatively short-term follow-up over 50% of our patients successfully have used tiered therapy from pacemaker-cardioverter defibrillators. This implies that these therapies protect selected young patients from sudden cardiac death, potentially greatly increasing their life span.

904-51 Long-term Outcome in Fontan Patients with Pacemakers

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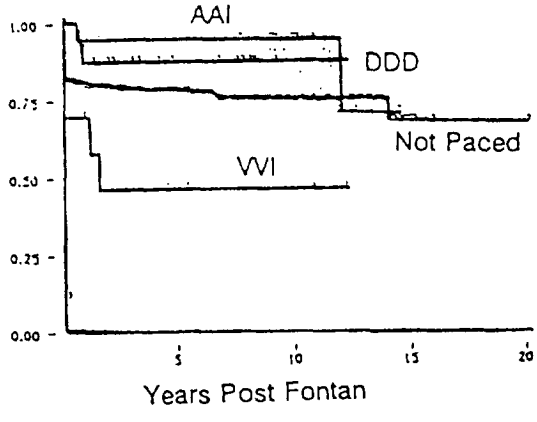
To further examine the outcome of Fontan pts with pacemakers we evaluated our database of 500 consecutive pts who underwent a Fontan operation at Children's Hospital in Boston between April, 1973 and August 1991 (mean follow up 3.5 ± 3.9 yrs, range 0–20.2 yrs). Pacemakers were implanted in 47



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pts (10%), 10 prior to Fontan, 3 at the time of Fontan, and 34 post Fontan. Indications for pacemaker implantation included sinus node dysfunction (SND) (n = 8), atrio-ventricular block (AVB) (n = 22), atrial flutter with (n = 13) or without (n = 2) coexisting SND, or SND and AVB (n = 2). Pacing modes included 9 with single chamber ventricular pacing (VVI), 19 with dual chamber pacing (DDD), and 19 with atrial pacing (AAI), of which 8 have antitachycardia pacing capability. Since 1983, most pts (90%) >2 yrs old with CHB received DDD pacing. The Kaplan-Meier survival curve compares the various pacing modes to non-paced pts.

Long term survival with a Fontan was significantly worse in pts with VVI pacemakers compared to the other groups, but those pts with VVI pacing were younger at the time of Fontan surgery or had earlier date of surgery. Outcome in AAI and DDD paced pts was similar to non-paced pts. "Physiologic" pacing (AAI or DDD) is preferable for single ventricle pts having a Fontan operation who require a pacemaker.

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904-52 Verapamil Therapy in Infants with Hypertrophic Cardiomyopathy

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The safety and efficacy of verapamil therapy in infants with hypertrophic cardiomyopathy (HCM) was prospectively evaluated in 17 children presenting at age 2 days to 1 year (median 3.5 months) with primary HCM (asymmetric or regional hypertrophy excluding infants of diabetic mothers, and metabolic disorders). Acute response to intravenous verapamil infusion was assessed during cardiac catheterization. Effects of chronic therapy were assessed by clinical, echocardiographic, and Holter monitor evaluation.

At presentation, 10 of 17 were symptomatic (7 congestive heart failure, 4 cyanosis, 1 arrhythmia, 1 apnea). Three cases were diagnosed on prenatal ultrasound. There was a positive family history in 7/15, 16/17 had murmurs, all had abnormal ECGs, 6 had weight >5%ile, and 2 had Noonan's syndrome.

Acute verapamil infusion was well tolerated by all, without adverse electrophysiologic effects. Hemodynamic effects of verapamil were consistent with a negative inotropic effect with a significant fall in cardiac index (4.6 ± 1.2 to 4.0 ± 0.7 l/min/m^2), no change in end-diastolic pressure (15 ± 8 mmHg before to 15 ± 6 mmHg after), and a small fall in systolic blood pressure (mean −7 mmHg). In the 8 patients with LV outflow obstruction, the LV to aortic gradient decreased by −18 mmHg mean (range +1 to −45). Followup is not available in 1 of 17 patients. Two patients underwent surgery for subaortic stenosis. During 1 to 116 months (mean 32 months) of oral verapamil therapy, two patients died (one sudden, one after cardiac transplant). The sudden death occurred during Holter monitoring and appeared unrelated to verapamil. Complete heart block was seen transiently in one but other significant electrophysiologic side effects were not observed. Among the 14 survivors with followup, 6 are clinically and echocardiographically improved, 8 are stable, and none have had clinical deterioration or worsening disease on echocardiography. Echocardiographically, LV outflow obstruction decreased in 8/10 patients, remained unchanged in 1, and increased in the patient who subsequently underwent cardiac transplantation. Two patients have had complete resolution of hypertrophy on echocardiogram.

We found that verapamil was well tolerated in infants with HCM without evidence of adverse effects and outcome was considerably better in these patients compared with prior reports. Resolution of hypertrophy in some patients suggests that either reversal of the phenotypic expression of this disorder is possible or else clinical and echocardiographic criteria may result in misdiagnosis.

904-53 Left Ventricular Structure and Function Eleven Years After Doxorubicin Treatment for Childhood Leukemia: Is this a Restrictive Cardiomyopathic Process?

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We previously characterized the progressive abnormalities of left ventricular (LV) structure and function in 115 survivors of childhood acute lymphoblastic leukemia 6.4 years (mean) after completion of doxorubicin therapy. 65% of patients having received >228 mg/m^2 of doxorubicin had either increased afterload or decreased contractility, or both. Progressive increases in afterload due to a thin LV wall appeared related to an inability to increase LV mass in response to somatic growth. Of these 115 patients, 36 have died, relapsed, or are no longer followed at our institution. 8 patients had both early and late congestive heart failure. Findings noted in patients with late heart failure included normal LV size in the setting of a mean LV fractional shortening <10%, as well as frequent pulmonary hypertension, increased pulmonary resistance, and left atrial dilation. We wished to characterize the late outcome of doxorubicin cardiotoxicity by reviewing the long-term echocardiographic findings of this cohort. The remaining 79 of the original 115 patients have had echocardiographic follow-up 11.1 years (mean) after completing doxorubicin. Changes in LV structure and function adjusted for age or body surface area at 3 time points were compared: prior to doxorubicin (A), 6.4 years after doxorubicin (B), and 11.1 years after doxorubicin (C). LV fractional shortening fell from normal at A to significantly depressed at B, and remained stable but depressed at C. LV mass was normal for body surface area at A, below normal at B, and continued to become more abnormal at C (p = 0.001). LV wall thickness was normal for body surface area at A, below normal at B, and continued to become more abnormal at C (p = 0.03). LV end-diastolic dimension was normal for body surface area at A and B, but during the subsequent 4.7 years, a significant fall in LV end-diastolic dimension for body surface area occurred (p = 0.03). LV afterload increased significantly from A to B, but remained elevated without further increase at C. LV contractility remained normal at all 3 times. In conclusion, the fall in LV dimension to subnormal levels with increasing follow-up, in spite of a reduced mass:volume ratio, is the pattern seen in restrictive cardiomyopathy with a loss of the capacity of the LV to dilate. Both the echocardiographic and clinical findings suggest that the moderate to long term course of children treated with boluses of doxorubicin may follow the clinical findings of a restrictive cardiomyopathic process with decreased LV compliance and a small thin-walled LV.

905 Clinical Utility of Pharmacologic Stress Echo Cardiography

Monday, March 20, 1995, Noon–2:00 p.m.
Ernest N. Morial Convention Center, Hall E
Presentation Hour: 1:00 p.m.–2:00 p.m.

905-54 Intercenter Agreement in Interpretation of Dobutamine Stress Echocardiograms. A Multicenter Trial

Rainer Hoffmann [1], Harald Lethen [1], Thomas Marwick [2], Mariarosaria Arnese [3], Paolo Fioretti [3], Alessandro Pingitore [4], Eugenio Picano [4], Thomas Buck [5], Raimund Erbel [5], Frank A. Flachskampf [1]. [1] *Med. Clinic I, Aachen, FRG;* [2] *Cleveland Clinic Foundation, USA;* [3] *Thoraxcenter Rotterdam, Netherlands;* [4] *University of Pisa, Italy;* [5] *Med. Clinic, Essen, FRG*

To assess the intercenter agreement in the interpretation of dobutamine stress echocardiograms (DSE), 5 experienced centers were each asked to submit 30 DSE (dobutamine up to 40 mcg/kg/min and atropine up to 1 mg) performed on patients (pts) with suspected coronary artery disease (CAD) undergoing coronary angiography. Thus, a total of 150 DSE were interpreted by each center unaware of any other patient data apart from the echos (no data on maximal dosage and reason for termination of DSE). No case was excluded due to low image quality or inadequate stress level. Data acquisition was performed in quad-screen format by 3 centers and on video tape by 2 centers.

Coronary angiography proved CAD in 95 pts (>50% diameter stenosis). Agreement in specific segments ranged from 84% to 97%, being highest for the basal anteroseptal segment and lowest for the basal inferior segment. On a patient basis, 4/5 or 5/5 of the five centers agreed on DSE abnormality or normality in 73%. Agreement was higher in pts with no CAD (82%; n = 55 pts) and in pts with 3 vessel disease (100%; n = 14 pts), and lower in pts with 1 and 2 vessel disease (61 and 68%, respectively).

Conclusion: Although there was heterogeneity in data acquisition, overall intercenter agreement in interpretation of stress echocardiograms is acceptable and comparable to similar data from perfusion scintigraphy. To increase intercenter agreement, a better standardization in image acquisition and reading criteria of stress echocardiography is warranted.

905-55 The Prognostic Value of Pharmacological Stress-Echocardiography: Experience in a Primary Care Cardiology Center

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Aim of the study was to evaluate the value of pharmacological stress echocardiography for risk stratification and clinical decision making in a primary care cardiology center.

A population of 526 patients (270 males and 256 females, age 60 ± 10 year, mean ± SD) with evidence, or suspected coronary artery disease were studied by pharmacological stress-echo. Patients underwent either Dipyridamole stress-echo (DIP) up to 0.84 mg/kg, or Dobutamine stress-echo (DOB) up to 40 μg/kg/min. Criteria for positivity of each test were an increase in wall motion score index during stress compared to pre-test, in a 16 segments model of left ventricle, each segment ranging from 1 = normal to 4 = diskinetic. DIP was performed in 398 patients and DOB in 128 patients. DIP was positive in